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A CASE OF ADIPOSIS DOLOROSA, WITH NECROPSY.*

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G. L., a white woman thirty-six years old, was brought to the Medico-Chirurgical Hospital in February of this year in semi-coma and unaccompanied by any one who could give an account of her illness.

She was very fat, weighing probably three hundred pounds. She was not, however, simply obese, because in addition to the uniform general increase in the sub-cutaneous fat there were great pendulous masses on the upper arms, the thighs and the abdomen which gave the sensation on palpation of a bundle of worms described by Dercum, and which were very sensitive to pressure, not only in the region of nerve trunks, but everywhere. The skin was not thickened nor adherent to the underlying tissue. In the skin over the masses there were many white, glistening superficial scars resembling those seen on the abdomen in women who have borne children. The hair, nails, and joints showed no trophic changes. There was no serous edema, no mucous edema, and no sign of acromegaly. The neck was so fat that it was impossible to palpate the thyroid gland.

She was, on admission, in extreme hebetude. She could with great difficulty be roused, but only for a moment. She answered questions, but only by single words, or if she attempted a sentence soon became confused and then silent and drowsy. She could give no account of her illness save to say that she had headache, felt sick, and could not walk, or see except dimly. Speech was slow, but not aphasic ncr inarticulate. She could not move either the arms or legs. She could flex and extend the fingers slightly, the toes not at all. She moved the head slowly and weakly, and swallowed what was put in her mouth, but made no attempt to chew. Sensory examination was difficult because of her mental dullness. Whether touch was felt could not be determined. Pain sense was preserved, and in the fatty masses there was hyperalgesia. The knee-jerks were very much diminished. The biceps tendon, plantar, jaw and abdominal jerks were absent. The condition of the muscle-jerks could not be determined on account of the fat overlying them. Vision was very bad, and there was marked optic neuritis. The thoracic and abdominal organs were normal. The urine at the first examination did not contain albumin, sugar, or tube casts, but later casts and albumin were present. Several days later I obtained the following fragmentary history from her brother: She had always been well till about two years ago, when she began to be heavy and dull, and changed from an active woman into a lethargic one. She began to grow fat several years before that. About six months before coming to the hospital she began to complain of weakness in walking. The difficulty slowly increased, and two weeks before admission she suddenly became palsied on the right side for a few hours. She then rapidly lost power in both legs and arms, and lost control of the bladder and rectum. She talked well until two weeks ago.

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She remained in the hospital several weeks, continuing semi-conscious, paying no attention to her bladder or rectum, speaking only a word or two now and then, never moving the arms or legs, and swallowing liquid food without trouble; then the coma deepened, and she died with edema of the lungs and acute Bright's disease. At necropsy there was found a tumor of the pituitary body about as large as a walnut, involving the optic chiasm and penetrating upward into the ventricles. There was quite marked internal hydrocephalus. The thyroid gland was normal in size, and contained a concretion about as large as a small chestnut. The lungs were edematous. The ovaries were small and hard.

My friend, Dr. D. J. McCarthy, hardened, sectioned, and examined pieces from the various organs. The fat revealed, on gross examination, a peculiar, fibrous character, it being firmer than normal subcutaneous fat, and showing here and there in encapsulated areas softer and less yellow in color. These isolated areas of fat varied in size from a pea to a hazelnut, and were surrounded by a distinct fibrous capsule. There appeared to be no connection between this fat and the firmer fat imbedded in the fibrous structure surrounding the fatty nodes. These nodules were not immediately beneath the skin, but deeper in the subcutaneous fat, and could not have been distinguished by palpation. On microscopic examination the firmer masses treated with connective tissue stains revealed a marked increase of this tissue, whereas the fatty nodule was normal in this regard, but an artery which ran through was distended and its coats were thickened. We were unable to find any nerves in the fatty nodules, but in the surrounding fat and in the underlying muscles the smaller nerve branches showed a high grade of interstitial neuritis. In the muscles this was associated with a granular fatty degeneration of the muscle fibers, the granules staining black with osmic acid. The interstitial changes in the fat nerves were more marked. The nerve fibers, when present, were few in number and surrounded by a dense connective tissue. In the nerve filaments in the muscle the connective tissue was decidedly increased, but there were more nerve fibers present than in the fat nerves.

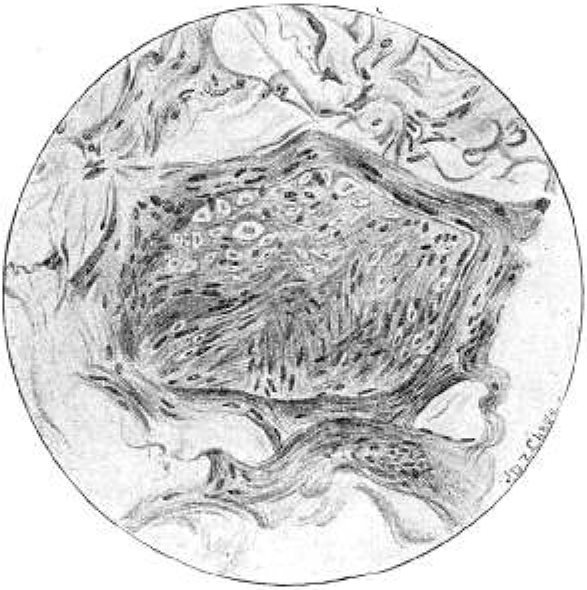

Fig. I.—Nerve twig in subcutaneous fat, showing high-grade interstitial neuritis. Oc. 3; ob. 7.

The ulnar, median, sciatic, peroneal and tibial nerves were examined. The Marchi, Weigert and carmine stains revealed no degeneration. In the sciatic nerves fewer nerve fibers were present than usual, and there was an infiltration of fat, most marked in the central areas, which gave to the fresh nerve a semi-gelatinous consistence.

The Brain.—A glioma the size of a walnut occupied the seat of the pituitary body and embraced the optic chiasm and nerves. The tumor extended into the interior of the brain, grew through the aqueduct of Sylvius, completely filling it and infiltrating its walls to a slight degree, and filled up the upper area of the fourth venticle. There was marked internal hydrocephalus.

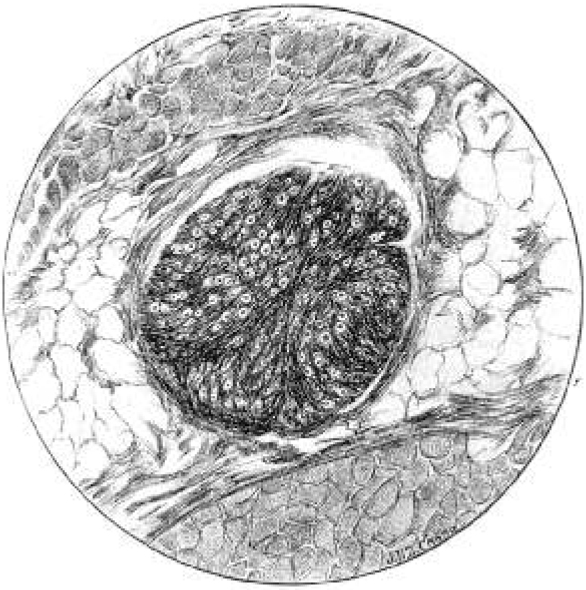

Fig. 11.—Nerve twig within the muscle, showing interstitial neuritis. Oc. 1; ob. 7.

The spinal cord showed a considerable grade of hydromyelia, extending down to the dorsal region. The external lateral group of cells of the anterior horn showed a light grade of chromatolysis.

In the right lobe of the thyroid gland there was a spherical concretion as large as a hickory nut. The gland was normal in size. On microscopic examination colloid degeneration with atrophy and absence of the secreting cells in many acini was found. There were many areas of small round cell infiltration and indications of an active inflammatory process were present everywhere. The muscles showed marked degeneration with the osmic acid method. It was probably secondary to degeneration of the nerves, as a medium grade of interstitial neuritis of the intramuscular nerve bundles was present.

There was acute parenchymatous nephritis with slight low-grade interstitial change. There was marked fatty infiltration of the liver.

The ovaries were examined by Dr. H. L. Williams, who reported as follows: "Surface smooth. No inflammation apparent. Tunica albuginea appears very much thickened, sclerotic and hyaline. The deeper tissue of the stroma is dense and contains large numbers of primordial Graafian follicles, and in the connective tissue beneath the ovarian stroma are large numbers of blood vessels with well-developed muscular walls. Scattered here and there through the stroma are a number of corpora fibrosa and also a few small cystic Graafian follicles. Projecting from a surface in one portion is a small body the size of a split pea, the tissue of which is identical with that of the ovarian stroma with which it is directly connected. Its surface is smooth, but inflamed, and it contains, a short distance below the surface, a number of undeveloped Graafian follicles, and just at the surface a small cystic follicle. The specimen is a small, typically sclerotic ovary."

Any one who had seen the original patients of Dercum could have made the diagnosis of adiposis dolorosa in this case with certainty and without difficulty. The picture was classical. But just as certainly there was some organic brain disease in addition. Hebetude and, indeed, continuous stupor are not uncommon in the last stages of adiposis dolorosa, but transient hemiplegia, a rapidly oncoming palsy of all four extremities, and optic neuritis cannot be caused by it. The symptoms showed that there was local brain disease of such a nature as to cause variations in the intra-cranial pressure, but it was impossible at the time I saw the patient to determine the situation of the lesion. Post mortem it was found in the pituitary body.

The case is of interest, because it throws some light upon the morbid anatomy of the disease, or at least shows what lesions are found. I know of no other necropsy in this disease, save the one which Dr. Dercum reports at this meeting. In general, his and my findings are the same—in both there is disease of the thyroid gland and of the finer nerve branches. But notwithstanding this agreement it is far too soon to have any fixed opinion as to the causation of adiposis dolorosa, or as to what organ is primarily at fault. The whole question is still in the region of hypothesis. Many more cases must be studied in life and death before safe conclusions can be drawn. Meanwhile in the present state of knowledge the thyroid gland is naturally first thought of as the organ disease of which would be apt to produce such symptoms. That disease of the thyroid gland profoundly affects nutrition is proven by myxedema and cretinism. The opinion that it may cause adiposis dolorosa rests rather on analogy than proof. Thyroidal concretions are not infrequently found accidentally post mortem and surely frequently do not cause symptoms of any kind. Much of the disease in the thyroid in my case was acute and hence could not have caused a condition which had begun years before. There were some pathological changes of long standing, however, and they may have had some relation with the adiposis dolorosa.

The most striking lesion was the tumor of the pituitary body. It completely destroyed all the glanular structure in that organ. This should have produced, according to a quite widely prevalent view, and according to my own published opinion, acromegaly. It did not. Whether the disease in it was a mere coincidence, or whether there was any causal relation between the tumor and the adiposis cannot be decided. It probably was coincidental, because in no other case has there been any evidence that the two conditions were associated—in no other case have there been present the symptoms of interference with the functions of the optic chiasm or the general symptoms of brain tumor. There may, however, in the cases studied only clinically have been atrophic disease of the pituitary body, and this would not cause symptoms which would attract attention to it. One thinks of the pituitary body only when there is acromegaly or the characteristic hemianopsia. Post mortem it is neglected unless there is some very patent lesion.

The ovaries are interesting. They were sclerosed and not performing their great function. Ovaries and testicles have something to do with fat formation. Spayed or castrated animals are almost always fat. Eunuchs are most often obese. Women at the menopause often put on much flesh. It is a physiological incident. Artificially induced menopause is often followed by obesity. But all these conditions are mere fatness—a very different thing from adiposis dolorosa. At present we can only say that adiposis dolorosa is a clinical entity having definite signs and symptoms, but with an as yet unknown pathology.

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107 LA MYOCLONIE EPILEPTIQUE (Epileptic Myoclonus). Rabot (Thèse de Paris, 1899).

The author states that Prichard was the first to describe this association. Delasiauve and Herpin were familiar with the phenomenon, as were other epileptologists. It has often been confounded with other affections, as by Hammond. Russell Reynolds stated that motor troubles of some sort occur in three-quarters of all epileptics. Recent authorities are cited by name, including some not usually enumerated (Weiss, Orazzio).

Nothing is more variable than the symptomatology of myoclonus in epilepsy. Féré has described bizarre movements in deglutition, and in the action of the diaphragm, producing abnormal respiration. Rabot also includes phenomena reported by the present reviewer (Dide) as belonging to myoclonus. Whatever occurs interparoxysmally of the nature of sudden repeated movements, having something of the quality of an electric shock, belongs here in Rabot's opinion. Rabot states positively that myoclonus, as an occasional symptom of epilepsy, is *not* the same affection as paramyoclonus multiplex, fibrillary chorea, myotonia, electric chorea, tic, hemiathetosis, etc. He would prefer to call it *petit mal moteur*, a manifest phenomenon of epilepsy proper.

CLARK.